

PEDIATRIC CARDIOLOGY

Assessment of Right Ventricular Performance by Pulsed Doppler Echocardiography in Patients After Intraatrial Repair of Aortopulmonary Transposition in Infancy or Childhood

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The purpose of this study was to determine whether measurement of aortic blood flow velocity and acceleration by pulsed Doppler ultrasound can be used to assess the global performance of the systemic right ventricle noninvasively in young patients who have undergone intraatrial repair of aortopulmonary transposition. The effect of age at surgery on right ventricular performance in these patients was also studied. Pulsed Doppler velocity profiles of aortic blood flow were analyzed at a mean age of 5.5 years in 24 patients who had undergone intraatrial repair of aortopulmonary transposition either in early infancy (mean age 3.4 ± 1.3 weeks) or later (mean age 8.5 ± 6.5 months). Velocity and acceleration variables in these patients were compared with the same variables in 24 age-matched normal subjects and with the performance of their own right ventricle as assessed by two-dimensional echocardiographic measurement of ejection fraction.

The 12 patients who underwent early repair had a higher ejection fraction than did the 12 who underwent

later repair (mean \pm SD 0.60 ± 0.07 versus 0.42 ± 0.10 ; $p < 0.001$). Aortic flow velocity was similar in all patients. Aortic acceleration was normal in patients after early intraatrial repair of aortopulmonary transposition (20.8 ± 2.3 m/s²), but was abnormally slow in patients after late repair (11.1 ± 1.8 m/s²; $p < 0.001$), thus suggesting that the latter group had diminished right ventricular performance. Concomitantly, acceleration time and ratio of acceleration time to ejection time were increased in patients after late repair. All three acceleration variables were linearly correlated with right ventricular ejection fraction ($r = 0.72$, $r = 0.63$ and $r = 0.71$, respectively; $p < 0.001$).

These findings indicate that measurement of aortic blood flow acceleration by pulsed Doppler ultrasound can be used to assess right ventricular performance noninvasively in patients after intraatrial repair of aortopulmonary transposition. The data also suggest that early intraatrial repair may promote better right ventricular performance.

(J Am Coll Cardiol 1989;13:1578-85)

Long-term results of the intraatrial repair of aortopulmonary transposition depend to a great extent on the ability of the right ventricle to function as the systemic pumping chamber. Because several studies (1-4) have documented that dysfunction of the right ventricle may occur in children who have undergone such repair, assessment of right ventricular performance is important during follow-up evaluations of

these patients. The noninvasive methods used to assess right ventricular performance after intraatrial repair of aortopulmonary transposition have included two-dimensional echocardiography (5,6), radionuclide imaging (7,8), and nuclear magnetic resonance imaging (9). Although two-dimensional echocardiography seems best suited for serial comparisons in these patients, its accuracy may be limited because the right ventricle is frequently enlarged or irregular in shape and may have abnormal regional wall motion (10,11). In addition, outlining the area of the right ventricle is time-consuming and requires computer facilities.

A different noninvasive method that could be used to assess systemic right ventricular performance is pulsed Doppler ultrasound, which is used to measure aortic blood flow velocity and acceleration as indexes of global left ventricular performance in adults (12-16). This method is independent of geometric assumptions, less time-consuming and does not require computer facilities. However, obtaining

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Manuscript received October 21, 1988; revised manuscript received December 7, 1988, accepted December 29, 1988.

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accurate Doppler tracings of aortic blood flow could be difficult in children with aortopulmonary transposition, because their aorta lies anterior to its normal position. The present study was undertaken to determine whether pulsed Doppler measurement of aortic blood flow velocity and acceleration can be used to assess the global performance of the systemic right ventricle easily and accurately in young patients who have undergone intraatrial repair of aortopulmonary transposition. For this purpose, we compared pulsed Doppler ultrasound measurements in 24 young people after such repair with both Doppler measurements in normal subjects, and measurements of their own right ventricular ejection fraction determined by two-dimensional echocardiography. We also studied the effect of age at surgery on right ventricular performance in patients after intraatrial repair of aortopulmonary transposition.

Methods

Study patients. We studied 24 asymptomatic infants, children and young people aged 1 month to 18 years (mean age 5.5 years), who were consecutively referred for follow-up evaluation after intraatrial repair of aortopulmonary transposition of the great arteries. We also studied 24 age-matched normal subjects who were referred for echocardiographic evaluation because of a functional murmur and were found to have no cardiac disease. Their measurements were used for comparison with those of patients who had undergone intraatrial repair of aortopulmonary transposition. In addition, to ensure that any differences found among patients were not caused by differences in age, body surface area or heart rate, we assessed the effect of these three variables on the Doppler indexes in these normal subjects.

Patients who had undergone intraatrial repair of aortopulmonary transposition were classified into two groups according to their age at the time of repair. Twelve had been operated on in early infancy (mean age \pm SD 3.4 ± 1.3 weeks). Twelve other patients had undergone repair later in infancy (mean age 8.5 ± 6.5 months), and three of them had required reoperation for various complications. The interval between surgery and Doppler echocardiographic study ranged from 3 weeks to 17 years. The clinical data, surgical procedure (Mustard or Senning operation) and postoperative complications of these patients are shown in Table 1. All patients were in New York Heart Association functional class I and were taking no medication at the time of study; 20 had sinus rhythm, 2 had junctional rhythm and 2 had an implanted pacemaker. Because all echocardiographic and Doppler studies were performed for clinical indications, no institutional approval was obtained.

Echocardiographic and Doppler studies. Two-dimensional echocardiography and pulsed Doppler ultrasound studies were performed with an ATL Ultramark 8 sector

scanner (Advanced Technology Laboratory) with use of a 3 or 5 MHz mechanical transducer; this instrument displays the Doppler shift as velocity. The subjects were examined in the supine position when subcostal or suprasternal views were used and in the left lateral decubitus position when left parasternal and apical views were used. When sedation was required, chloral hydrate (75 mg/kg body weight [not >1.5 g]) was given. Two-dimensional imaging and Doppler velocity curves were recorded on 0.5 in. (1.27 cm) videotape simultaneously with an electrocardiogram; the display speed of the Doppler tracing was 100 mm/s.

Doppler velocity tracings of ascending aorta blood flow were recorded from a variety of views to obtain the highest possible velocity (17). Suprasternal, apical, high left parasternal and subcostal windows were used, and the sample volume was placed just distal to the level of the aortic valve. The velocity tracings with the highest velocity and the smallest spectral dispersion were selected for further analysis (Fig. 1). Because the angle of incidence was $<20^\circ$ in all recordings, flow velocities were not corrected for angle.

Analysis of echocardiographic and Doppler data. Off-line analysis of echocardiographic and Doppler data was performed with a microcomputer system (CAD 886; Microsonics). For echocardiographic evaluations, apical four chamber and parasternal short-axis views of the right ventricle at end-diastole and end-systole were digitized. From area outlines in these images, right ventricular end-diastolic volume (EDV) and end-systolic volume (ESV) were determined with use of a Simpson's rule biplane algorithm as previously described, in patients after the Mustard operation for aortopulmonary transposition (5). Ejection fraction (EF) was then calculated as $EF = (EDV - ESV)/EDV$. Because the biplane method disregards the infundibular part of the right ventricle, the echocardiographic measurements of right ventricular volumes were repeated with use of a modified area-length method that includes the infundibulum (6).

For pulsed Doppler evaluations, recordings of aortic blood flow were also digitized. The following variables were measured or calculated from these tracings: peak and mean aortic flow velocities, acceleration time, ejection time, mean aortic acceleration and ratio of acceleration time to ejection time (Fig. 1). Peak aortic flow velocity was measured at the center of the Doppler flow spectrum at the time of maximal velocity (17). Mean aortic flow velocity was calculated as the aortic flow velocity time integral divided by the ejection time: the flow velocity time integral was determined by integrating the area under the aortic Doppler velocity curve. Acceleration time (AT) and ejection time were measured from the onset of ejection to the point of peak velocity (PkV) and to the end of ejection, respectively. Mean aortic acceleration (A) was calculated as $A = PkV/AT$. Each Doppler measurement was repeated in three to five consecutive cardiac cycles and averaged.

Table 1. Clinical and Surgical Data and Right Ventricular Ejection Fraction in 24 Children After Atrial Repair of Aortopulmonary Transposition

Patient No.	Age (yr)	BSA (m ²)	Surgical Procedure	Operative Age (mo)	Postoperative Complication	RVEF
Early repair						
1	4	0.65	Senning	1	Mild LVOT obstruction	0.65
2	2.5	0.55	Senning	0.75	Endocarditis	0.72
3	8	0.92	Mustard	1.25	Pacemaker	0.69
4	4	0.67	Senning	0.4	None	0.63
5	0.1	0.20	Senning	0.5	None	0.62
6	0.2	0.33	Senning	1	None	0.64
7	1.5	0.50	Senning	1	None	0.54
8	4	0.70	Senning	1.5	None	0.51
9	0.5	0.44	Senning	0.5	Mild SVC obstruction	0.49
10	1.5	0.45	Senning	1	None	0.51
11	0.1	0.22	Senning	0.75	None	0.65
12	0.2	0.24	Senning	0.5	None	0.59
Mean	2.2	0.49		0.85		0.60
±SD	±2.3	±0.22		±0.34		±0.07
Late repair						
13	5	0.71	Mustard	6	Mild LVOT obstruction	0.27
14	4	0.72	Senning	5	Mild LVOT obstruction	0.39
15	5	0.75	Mustard	2 and 30	Baffle leak (reoperated)	0.53
16	3	0.60	Senning	2 and 18	LV-PA conduit (reoperated)	0.56
17	10	1.14	Mustard	6	None	0.44
18	11	1.20	Mustard	6	None	0.31
19	15	1.60	Mustard	18 and 24	PVR obstruction (reoperated)	0.52
20	18	1.79	Mustard	12	Junctional rhythm	—
21	7.5	0.80	Mustard	12	None	—
22	4	0.70	Senning	6	None	—
23	10	1.48	Mustard	4	Junctional rhythm	0.36
24	14	1.40	Mustard	24	Pacemaker	0.41
Mean	8.8*	1.07*		8.5*		0.42*
±SD	±4.7	±0.41		±6.5		±0.10

*p < 0.001 versus early repair. BSA = body surface area; LVOT = left ventricular outflow tract; LV-PA = left ventricle to pulmonary artery; PVR = pulmonary venous return; RVEF = right ventricular ejection fraction; SVC = systemic venous connection; — = not obtained.

Intraobserver and interobserver variability. To test the repeatability of the Doppler measurements of aortic flow velocity and acceleration, measurements were made in duplicate. To test their reproducibility, aortic flow velocity and acceleration were remeasured by a second investigator who was unaware of the other's results or of the right ventricular ejection fraction.

Statistical analysis. Adequate Doppler recordings of aortic flow were obtained in all subjects. However, in 3 of the 24 patients with aortopulmonary transposition who had an enlarged right ventricle, it was technically impossible to determine the right ventricular ejection fraction accurately by two-dimensional echocardiography.

One-way analysis of variance was used to compare Doppler ultrasonographic measurements of aortic flow velocity and acceleration in patients after intraatrial repair of aortopulmonary transposition with measurements in normal subjects. Linear regression analysis was used to compare

Doppler data with right ventricular ejection fraction in patients with aortopulmonary transposition; it was also used to compare the two methods used to determine right ventricular volumes and to study whether Doppler indexes are related to age, body surface area or heart rate in normal subjects.

Results

Doppler ultrasound findings in young normal subjects and patients with transposition (Table 2). Doppler measurements of aortic blood flow velocities were similar in all groups, but mean aortic acceleration was abnormally slow in patients who had had late intraatrial repair of aortopulmonary transposition (Fig. 2). Values of mean aortic acceleration in normal subjects and patients after early intraatrial repair (>15 m/s²) did not overlap the values of patients after late repair (<15 m/s²) (Fig. 3). Similarly, aortic acceleration time

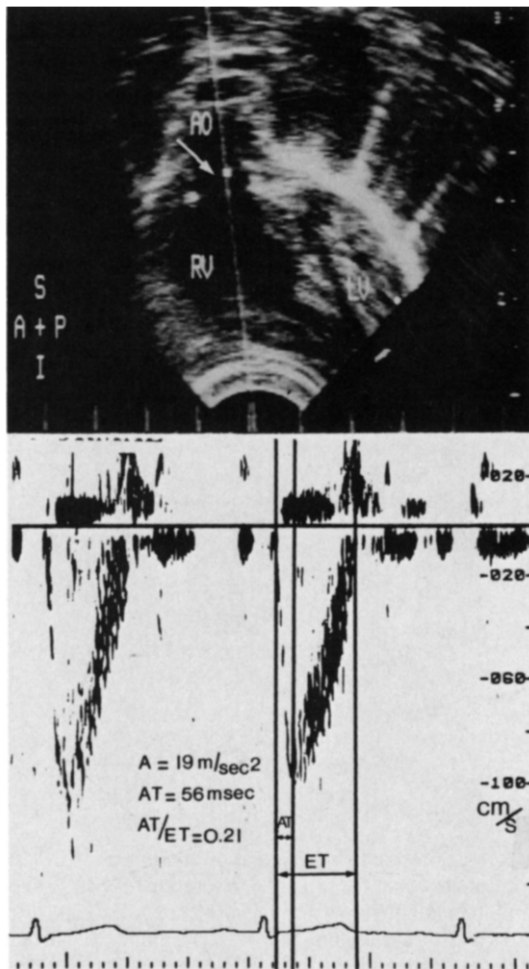


Figure 1. Pulsed Doppler echocardiographic assessment of right ventricular performance. **Top,** Subcostal imaging in a patient with aortopulmonary transposition after intraatrial repair, demonstrating the ascending aorta (AO) located anteriorly (A) and arising from the right ventricle (RV). The Doppler sample volume (arrow) is placed just above the aortic valve. **Bottom,** Pulsed Doppler tracing of the aortic blood flow recorded in a normal subject. The way acceleration time (AT) and ejection time (ET) were measured is demonstrated, and the values for AT, AT/ET and mean aortic acceleration ($A = \text{peak velocity}/AT$) are given. Peak and mean aortic flow velocity values in this subject were 96 and 61.4 cm/s, respectively. I = inferior; LV = left ventricle; P = posterior; S = superior.

was longer and the ratio of acceleration time to ejection time was greater in patients after late intraatrial repair of aortopulmonary transposition than in normal subjects and in patients after early repair. For the latter indexes, however, some values in normal subjects and patients after early intraatrial repair overlapped those in patients after later repair (Fig. 3).

Doppler ultrasound findings and right ventricular ejection fraction. The two methods used to assess right ventricular ejection fraction gave very similar results and correlated well with each other (Fig. 4). The results obtained with the biplane method (5) are reported and were used for further comparisons. Right ventricular ejection fraction was significantly higher in patients after early repair (0.60 ± 0.07) than in patients after late repair (0.42 ± 0.10 , $p < 0.001$) (Table 1). Neither peak nor mean aortic velocity correlated with right ventricular ejection fraction. In contrast, mean aortic acceleration, acceleration time and the ratio of acceleration time to ejection time each correlated linearly with right ventricular ejection fraction (Fig. 5). The correlations of mean aortic acceleration and the ratio of acceleration time to ejection time with ejection fraction were stronger than the correlation of acceleration time with ejection fraction.

Doppler data and age, body surface area and heart rate. Age, body surface area and heart rate differed between the two groups of patients with early and late repair of aortopulmonary transposition (Tables 1 and 2). However, in the 24 normal subjects studied, none of these variables correlated with peak or mean aortic flow velocity, mean aortic acceleration, or ratio of acceleration time to ejection time. Only acceleration time increased slightly with age ($r = 0.43$, $p < 0.05$) and with body surface area ($r = 0.50$, $p < 0.02$).

Intraobserver and interobserver variability. The measurements of aortic flow velocity, acceleration time and ejection time were repeatable and reproducible. Intra- and interobserver variability was $<5\%$.

Discussion

Assessment of right ventricular performance. The present study shows that measurements of acceleration variables of

Table 2. Doppler Measurements of Aortic Blood Flow in 24 Normal Subjects and 24 Infants, Children and Young People After Repair of Aortopulmonary Transposition

Group	Heart Rate (beats/min)	Aortic Flow Velocity (cm/s)				
		Peak	Mean	A (m/s^2)	AT (ms)	AT/ET
Normal (n = 24)	104 ± 32	105 ± 19	63 ± 14	21.1 ± 3.6	52 ± 15	0.21 ± 0.06
Early repair (n = 12)	108 ± 26	102 ± 16	60 ± 14	20.8 ± 2.3	50 ± 12	0.22 ± 0.03
Late repair (n = 12)	$75^* \pm 15$	103 ± 8	61 ± 11	$11.1^* \pm 1.8$	92 ± 12	$0.32^* \pm 0.04$

* $p < 0.001$ versus normal children. Values are expressed as mean \pm SD. A = mean aortic flow acceleration; AT = acceleration time; ET = ejection time.

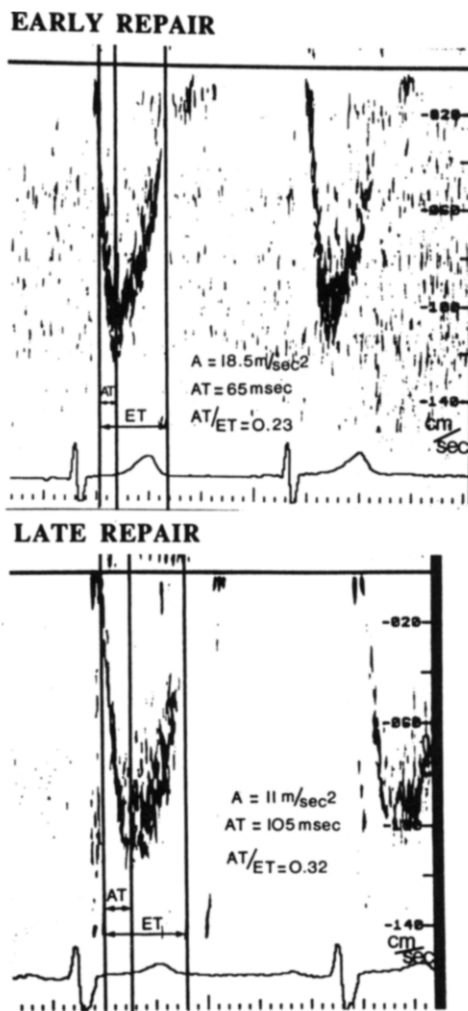


Figure 2. Pulsed Doppler recordings of the aortic flow velocity curve in Patient 8, a 4 year old child, after early intraatrial repair of aortopulmonary transposition (**top**) and in Patient 19, a 15 year old girl, after late intraatrial repair of aortopulmonary transposition (**bottom**). Note the differences in aortic acceleration (A), acceleration time (AT) and the ratio of acceleration time to ejection time (ET).

aortic blood flow by pulsed Doppler ultrasound can be used to assess global right ventricular performance easily and accurately in young patients after intraatrial repair of aortopulmonary transposition. Measurements of aortic blood flow velocity, however, do not indicate right ventricular performance accurately. The pulsed Doppler method is easy to use because it requires only a good aortic flow tracing in which three simple measurements are made; such a tracing was readily recorded in all our patients. In contrast, two-dimensional echocardiographic assessment of right ventricular performance requires imaging in several views and time-consuming area-outlining of the ventricle; appropriate imaging may sometimes be impossible, as was the case in three of our patients. The accuracy of the pulsed Doppler

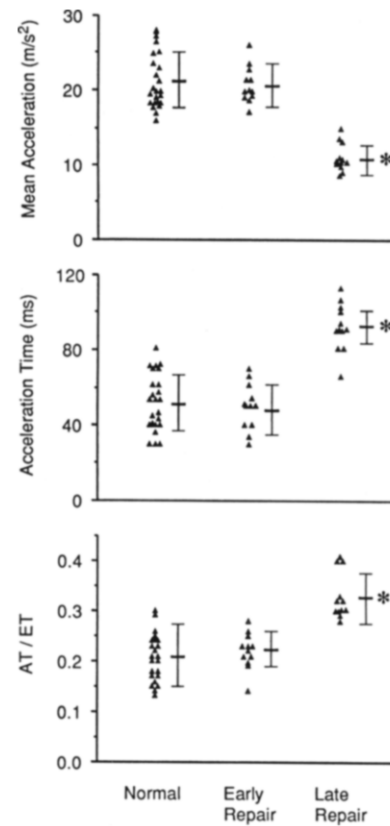
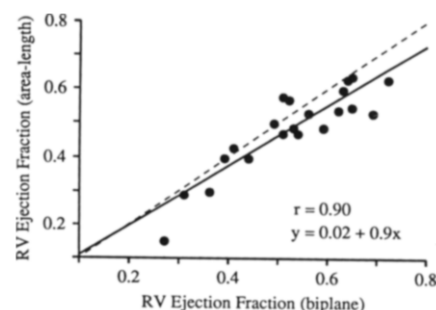


Figure 3. Mean aortic flow acceleration (**top**), aortic flow acceleration time (**middle**), and the ratio of acceleration time (AT) to ejection time (ET) (**bottom**) in young normal subjects and patients after early and late repair of aortopulmonary transposition. *Significantly different from normal subjects ($p < 0.001$).

method is shown by our finding of a linear correlation for all acceleration variables (mean aortic acceleration, acceleration time and ratio of acceleration time to ejection time) with right ventricular ejection fraction; the acceleration variables allowed us to distinguish clearly between patients with

Figure 4. Linear correlation between right ventricular (RV) ejection fraction derived from volume calculations by a biplane Simpson's rule algorithm (5) (x axis) and by a modified area-length method (6) (y axis). The **dotted line** is the line of identity; the **solid line** is the regression line.



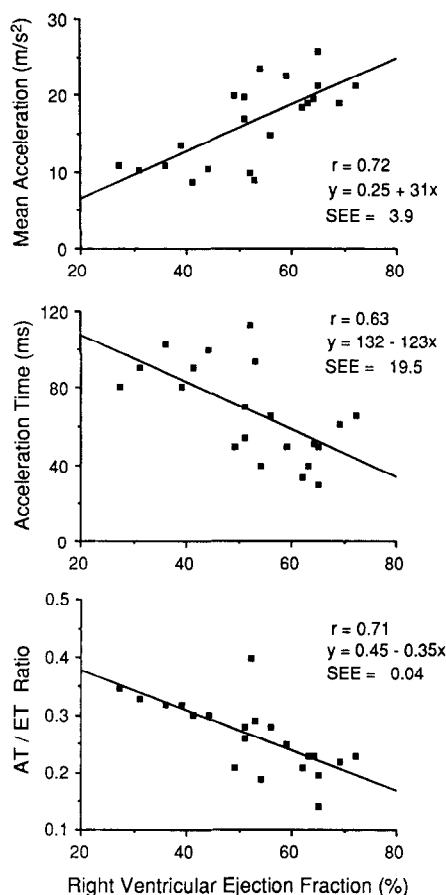


Figure 5. Linear correlations between right ventricular ejection fraction determined by two-dimensional echocardiography and mean aortic flow acceleration (**top**), acceleration time (**middle**) and the ratio of acceleration time (AT) to ejection time (ET) (**bottom**) measured by pulsed Doppler ultrasound in 21 young patients after intraatrial repair of aortopulmonary transposition.

normal and those with impaired right ventricular performance. In contrast, we found no correlation between peak or mean aortic flow velocity and ejection fraction.

Support for the use of pulsed Doppler ultrasound to assess right ventricular performance in infants, children and young people after intraatrial repair of aortopulmonary transposition is found in the similarity of our findings in these patients, for whom the right ventricle is the systemic pumping chamber, to findings in adults with normally related great arteries and a systemic left ventricle. Specifically, strong correlations between aortic flow acceleration and left ventricular ejection fraction have been reported (13-16) in adult patients with normal or reduced left ventricular performance due to dilated cardiomyopathy or ischemic heart disease. Similarly, aortic flow acceleration variables correlated with the right ventricular ejection fraction in our young patients. Aortic flow acceleration variables may be particularly useful to assess right ventricular performance in patients with aortopulmonary transposition because, as has

been shown in normal adults (15), they reflect changes in the inotropic state rather than changes in the loading conditions of the heart.

The finding that aortic flow velocity is not a useful indicator of right ventricular performance in our patients is not surprising. Although aortic peak and mean velocities have been shown to decrease in adults when severe left ventricular dysfunction is present, in such patients these variables correlated less well with ejection fraction than did aortic acceleration (14,16). The reason for the lack of correlation between aortic flow velocity and right ventricular ejection fraction in our patients may be that aortic peak and mean velocities are related to stroke volume (14,18,19); the normal peak and mean aortic velocities in our asymptomatic patients with transposition suggest that they had normal stroke volume and cardiac output at rest. Normal cardiac output at rest, however, does not necessarily indicate normal right ventricular performance in these patients (1,10,20). Probably, more severe dysfunction of the systemic ventricle leading to clinical symptoms such as congestive heart failure must occur before decreased aortic flow velocities can be observed, as was reported in adult patients with poor left ventricular performance (14,16). Thus, mild to moderate right ventricular dysfunction, which was present in some of our asymptomatic patients with aortopulmonary transposition, would be undetectable by Doppler measurement of aortic flow velocity.

Validation of the method. It was not possible to validate the pulsed Doppler assessment of right ventricular performance with invasive methods such as ejection fraction calculated from angiography, because most of our patients had not undergone recatheterization at the time of this study. However, echocardiographic determinations of right ventricular ejection fraction have been shown to correlate very well with angiographic determinations of ejection fraction when used in patients after intraatrial repair of aortopulmonary transposition (5,6). We therefore validated our pulsed Doppler measurements against right ventricular ejection fraction determined by two-dimensional echocardiography. For this validation we chose to use the values calculated by the biplane method (5) because it may better take into account any irregularities of the shape that occur in the short-axis dimension of patients with a systemic right ventricle. Although the biplane method disregards the infundibular part of the right ventricle, the infundibular contribution to right ventricular end-diastolic volume and to stroke volume is small and does not greatly influence the estimate of ejection fraction. In addition, ejection fraction value determined by the area-length method, which includes the infundibular part of the right ventricle, correlated excellently with those determined by the biplane method.

Other echocardiographic methods, using subcostal imaging of the right ventricle, have recently been proposed (11) for evaluating right ventricular performance in patients with

aortopulmonary transposition. Although the subcostal approach may be better suited for the study of small infants, it has limited use in older patients, in whom parasternal and apical images can be recorded with greater frequency.

Early versus late repair. Our findings also suggest that age at surgery affects right ventricular performance during follow-up of young patients who have undergone intraatrial repair of aortopulmonary transposition. In our study groups, right ventricular ejection fraction and mean aortic acceleration were significantly less in patients who underwent late than in those who underwent early repair, and acceleration time and the ratio of acceleration time to ejection time were significantly greater in the former. These results are consistent with previous reports (1,2) that right ventricular ejection fraction was reduced in asymptomatic patients who underwent intraatrial repair late in infancy or in childhood. It is conceivable that the normal values in patients who underwent early repair may result from less prolonged hypoxia in the ventricle, which is not intended to carry the burden of a systemic pumping chamber. It is also possible, however, that reduced right ventricular performance in patients operated on later results from less optimal experience with this operation during the earlier years when the operation was performed or from a longer course of the disease. It remains to be determined whether all our patients with normal right ventricular performance will keep this functional status, because they have been followed up for only a limited period in relation to their expected life span.

We can rule out the possibility that the different values of aortic acceleration variables in the study groups with earlier versus late repair resulted from differences in age, body surface area or heart rate. Although aortic acceleration may decrease with age (21,22), in the limited age range of our normal subjects we did not find any significant relation between acceleration or ratio of acceleration time to ejection time and age, body surface area or heart rate; we found only an increase in acceleration time with increasing age or body surface area. However, the absolute values of mean acceleration of aortic flow measured in our group of normal subjects and in our group of young patients after early repair of transposition were higher than those reported in normal adults (14) (15.5 to 28 and 17.1 to 26 m/s² versus 7.3 to 13.1 m/s²). Although this difference may reflect decreasing aortic acceleration with age (21,22), it might also be explained in part by different methods used to measure velocity from the Doppler recordings (14).

Conclusions. Measurement of aortic flow acceleration by pulsed Doppler ultrasound is an easy and accurate method for assessing right ventricular performance noninvasively in young patients after intraatrial repair of aortopulmonary transposition. It seems possible that the Doppler measurement of aortic flow could also be used to assess systemic ventricular performance in patients after the arterial switch operation for aortopulmonary transposition as well as in

patients with other forms of complex congenital heart disease such as congenitally corrected aortopulmonary transposition or univentricular atrioventricular connection. This study also suggests that, if intraatrial repair of aortopulmonary transposition is considered, it is probably optimally performed during the first few weeks of life to preserve normal systemic right ventricular performance after repair.

We thank Mimi Zeiger for editorial assistance.

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